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Follow-Up of Aortic Coarctation Repair in Neonates

Michael D. Puchalski, MD, Richard V. Williams, MD, John A. Hawkins, MD, L. LuAnn Minich, MD, Lloyd Y. Tani, MD

Salt Lake City, Utah

OBJECTIVES The purposes of this study were to assess the growth of left heart (LH) structures, to evaluate midterm outcomes, and to define echocardiographic parameters predictive of increased risk of re-intervention in patients born with aortic coarctation and hypoplasia of LH structures that underwent neonatal coarctation repair.

BACKGROUND Neonatal coarctation is often associated with hypoplasia of LH structures. Although previous studies have shown that coarctation repair can be performed with good results in these neonates, there are little data regarding growth of the LH structures or outcomes in these patients.

METHODS Patients with isolated coarctation and at least one hypoplastic LH valve (mitral or aortic Z-score <-2) who underwent a neonatal coarctation repair were identified. Clinic charts and the latest echocardiograms were reviewed.

RESULTS All 55 patients were alive and well, and no patient had clinical evidence of mitral stenosis. Three patients (5%) required re-intervention. Thirty-eight patients had echocardiograms that demonstrated normal left ventricular (LV) size and function with a follow-up duration of 73 ± 19 months (range 3 to 9 years). Both mitral and aortic annulus Z-scores increased significantly: -3.1 ± 1.5 to -0.5 ± 1.6 ($p < 0.001$) and -3.5 ± 1.9 to 0.7 ± 1.6 ($p < 0.001$), respectively. Nine patients (24%) developed LV outflow tract obstruction by echocardiographic criteria.

CONCLUSIONS After neonatal coarctation repair with associated LH hypoplasia, LH structures increase substantially in size, and clinical outcomes are excellent at midterm follow-up. Despite initial annular hypoplasia, the need for intervention for mitral or aortic/subaortic stenosis is uncommon. (J Am Coll Cardiol 2004;44:188–91) © 2004 by the American College of Cardiology Foundation

Aortic coarctation is often associated with hypoplasia of other left heart (LH) structures including the mitral valve, left ventricle (LV), and aortic valve (1). When these structures are hypoplastic, determining the adequacy of the LH for supporting the systemic circulation is critical for deciding on the best treatment option. If the LH is too small to support the systemic circulation, the patient should undergo surgical palliation for a single ventricle or heart transplantation, thus eliminating the possibility of a biventricular repair. Extensive investigation has been done to determine the adequacy of LV size for a biventricular repair in patients with critical aortic stenosis (2–5), but few studies have focused on LH hypoplasia in patients with aortic coarctation. Previous studies have shown that neonates with coarctation and LH hypoplasia can undergo successful coarctation repair and that the sizes of LH structures are not predictive of successful immediate or short-term outcome (6–8). Despite this, other investigators have found age at surgery and aortic arch hypoplasia to be risk factors for recoarctation (9,10), but these reports fail to account for other factors that influence hemodynamics and affect long-term outcome. This study was undertaken to assess growth of the LH structures, to evaluate midterm outcomes, and to define echocardiographic parameters predictive of increased risk of re-intervention in patients born with aortic coarctation and hypoplasia of LH structures who underwent neonatal coarctation repair.

From the Primary Children's Medical Center, University of Utah, Salt Lake City, Utah.

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MATERIALS AND METHODS

Patients studied. All patients studied were recruited from the original cohort in the Tani et al. study (6). The following were the criteria for inclusion at presentation:

- an echocardiographic diagnosis of coarctation of the aorta between January 1, 1993, and April 30, 1997, leading to surgical repair at Primary Children's Medical Center within the first two months of life;
- patency of all four cardiac valves with a mitral or aortic valve Z-score <-2;
- normally related and concordantly connected arterial trunks;
- absence of a true atrial septal defect or anomalous pulmonary venous connection; and
- absence of significant valvar or subvalvar aortic stenosis (peak Doppler gradient <20 mm Hg).

The medical records were reviewed for current patient age, body surface area, date of surgery, follow-up duration, outcomes, need for re-intervention, and most recent echocardiogram. The Institutional Review Board at the University of Utah School of Medicine approved this study.

Echocardiography. Two-dimensional, M-mode, and Doppler echocardiography were performed using an Acuson 128 XP or Sequoia (Acuson, Mountain View, California),

Abbreviations and Acronyms
LH = left heart
LV = left ventricle/ventricular

or a Hewlett-Packard Sonos 5500 system (Agilent, Andover, Massachusetts) with images recorded on 0.5-inch videotape. In the original study, the dimensions of the aortic valve, aortic root, mitral valve, and transverse arch were measured. The LV end-diastolic area and dimension, LV ejection fraction and shortening fraction, long-axis ratio, and Rhodes score (2) were calculated. For this study, the most recent echocardiogram was reviewed and optimal images were selected for offline measurements using previously described techniques (11). The dimensions of the aortic valve annulus, aortic root, and the mitral valve annulus (antero-posterior diameter) were measured from the parasternal long-axis view. M-mode measurements of LV end-diastolic dimension and LV shortening fraction were obtained from the parasternal short-axis view. The lateral dimension of the mitral valve annulus and the LV ejection fraction (Simpson's method) were measured from the apical window. Z-scores were calculated using formulas that compared the measured cardiac structure with normal values for a given body size (12). A structure was defined as being hypoplastic if the Z-score was <-2.0. The transverse arch was measured between the left common carotid artery and the left subclavian artery from suprasternal notch or high parasternal images.

Statistical methods. Paired *t* tests were used to compare Z-scores before and after coarctation repair. Unpaired *t* tests were used to determine if echocardiographic parameters were associated with the need for re-intervention. A chi-square test was used to determine if a size cut-off was predictive of the need for re-intervention. All values are expressed as mean ± SD. Statistical significance was inferred when p was < 0.05. Statistical analyses were performed using SigmaStat (SPSS Inc., Chicago, Illinois).

RESULTS

Patients. Of the 63 patients in the original study, 55 (87%) had clinical data available, and 38 patients (60%) had a recent follow-up echocardiogram. The remaining eight patients were lost to follow-up. The mean duration of follow-up was 73 ± 19 months (range 3 to 9 years).

Echocardiographic data. For the group of 38 patients who had follow-up echocardiographic data, all 38 had normal LV end-diastolic dimensions (37 ± 4 mm), shortening fractions (39 ± 6%), and ejection fractions (65 ± 5%). In all 38 patients, both mitral (Fig. 1) and aortic (Fig. 2) Z-scores increased significantly. Of the 29 patients (76%) who had mitral valves that were hypoplastic at birth (Z-score <-2), only 6 (16%) remained hypoplastic at follow-up ($p < 0.001$). Of the 31 patients (82%) who had aortic valve hypoplasia at birth, only 2 (5%) remained hypoplastic at follow-up ($p < 0.001$).

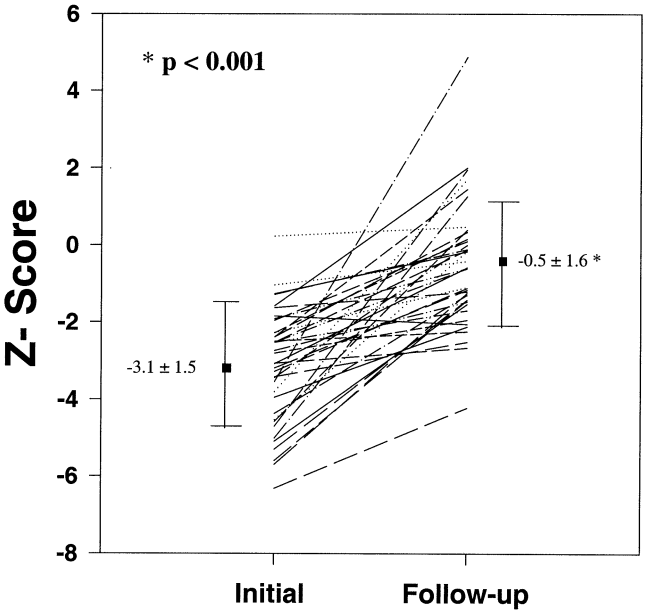

Figure 1. Initial and follow-up mitral valve Z-scores.

Outcomes and re-intervention. All 55 patients with available data were alive, and none was converted to single-ventricle physiology. Eleven patients (20%) required re-intervention: three (5%) for subaortic membrane resection, two (4%) for balloon aortic valvuloplasty (both with previous subaortic membrane resection), and nine (16%) for balloon dilation or surgery for recoarctation (one patient required all three procedures). Mitral valve Z-scores, aortic valve Z-scores, and transverse arch diameter at presentation were not different between patients requiring and those not requiring re-intervention (Table 1).

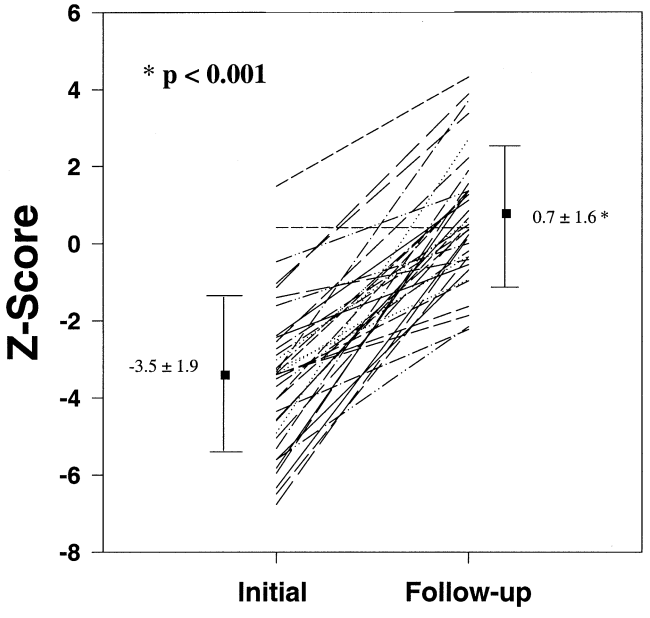

Figure 2. Initial and follow-up aortic valve Z-scores.

Table 1. Initial Left Heart Structure Size Versus Need for Re-Intervention

	Initial Mitral Annulus Z-score	Initial Aortic Annulus Z-score	Initial Transverse Arch Diameter (mm)
Re-intervention (n = 11)	−2.99 ± 1.82	−3.45 ± 2.5	3.96 ± 0.92
No re-intervention (n = 44)	−2.98 ± 1.59	−2.95 ± 1.99	3.94 ± 0.72
p Value	0.984	0.477	0.944

Of the 38 patients who had echocardiograms, 9 (24%) developed LV outflow tract obstruction, including 6 (16%) with isolated subaortic stenosis, 3 (8%) with isolated valvar aortic stenosis, and 2 (5%) with both. Although not clinically apparent, one patient had mild echocardiographic evidence of mitral stenosis (13).

DISCUSSION

Despite significant annular hypoplasia at presentation, the aortic and mitral valves of neonates undergoing aortic coarctation repair showed significant increases in their Z-scores on follow-up. In addition, midterm outcomes were excellent, and the need for re-intervention was uncommon.

Previous studies have suggested the possibility of postnatal growth of LH structures (2,8,14). Krauser et al. (15) studied infants with aortic coarctation and found that LV size appeared small preoperatively for two reasons: 1) compression of the LV by the dilated, pressure-loaded right ventricle; and 2) underfilling of the LV because of atrial-level left-to-right shunting. In an effort to determine the importance of preoperative hypoplasia of the LH structures, this study focused on the Z-scores of the diameters of the aortic and mitral valves and demonstrated a significant increase for both valves.

Several studies have shown that biventricular repair is possible with an aortic or mitral valve orifice diameter >2 standard deviations below normal. These studies, however, focused on early survival (6,7). Our study expanded on these data by showing that midterm survival and outcomes were excellent. All patients were alive, and none was converted to single-ventricle physiology, indicating the importance of considering growth potential in addition to other factors when determining the adequacy of the LV to support the systemic circulation.

Annular hypoplasia at presentation also failed to predict significant postoperative mitral, aortic, or subaortic stenosis. In fact, re-intervention for aortic/subaortic stenosis was uncommon. These findings conflict with the previous report of Levine et al. (1) in several important ways. They found 101 patients <3 months of age who presented with aortic coarctation and had a mean follow-up of 2.3 years. Their conclusion was that a mitral valve Z-score <−2 was associated with increased risk for obstruction at any level within the LH excluding recoarctation (up to 80% by 5 years). Our data showed that despite an average mitral valve Z-score of −3 at presentation, only nine patients (24%) subsequently developed aortic or subaortic stenosis with a mean follow-up of six years. Second, 17% of their patients developed LV outflow obstruction requiring re-intervention, compared with 5% in our series. Finally, Levine et al. (1) reported 11 patients (11%) who developed mitral stenosis, with 6 (6%) requiring intervention, whereas only 1 patient (2%) in our series developed echocardiographic mitral stenosis that was not clinically apparent. The reason for the differences in our findings is unknown, but this difference cannot be explained by the duration of follow-up, as it was almost four years longer in our study group.

The incidence of recoarctation in our study (16%) is similar to previous studies reporting recoarctation in 9% to 23% of patients (9,16–19). However, unlike previous studies (16), a transverse arch diameter of <3.5 mm did not significantly predict the need for re-intervention.

Study limitations. This study is limited by the retrospective design. A few patients (13%) were lost to follow-up. An additional 17 patients did not have follow-up echocardiograms, limiting the size of the sample used for assessing the growth of the mitral and aortic valves. However, the lack of a recent follow-up echocardiogram would suggest that the cardiologist felt these patients were clinically well and that a follow-up echocardiogram was not warranted.

Conclusions. After neonatal coarctation repair with associated LH hypoplasia, LH structures increase substantially in size, and clinical outcomes are excellent at midterm follow-up. Despite initial annular hypoplasia, the need for intervention for mitral or aortic/subaortic stenosis is uncommon.

Reprint requests and correspondence: Dr. Michael D. Puchalski, University of Utah, Primary Children's Medical Center, 100 North Medical Drive, Salt Lake City, Utah 84113. E-mail: pcmpucha@ihc.com.

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